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Case Series

Intestinal Pneumatosis, Chronic Bronchopneumopathy and Autoimmune Diseases: A Rare Finding with Variable Clinical Onset. A Case-Series

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Abstract

Intestinal Pneumatosis (PI) has been reported to be associated with many gastrointestinal disorders including obstruction, ischemia and infection [1]. PI has also been described in patients with Chronic Obstructive Pulmonary Disease (COPD), leukaemia, connective tissue disorders, organ transplantation and various immunodeficiency conditions [2-5]. There is a strong link with autoimmune diseases such as Sjögren Syndrome and Systemic Sclerosis [6]. PI is characterized by gas-filled cystic lesions within the wall of the bowel and can have a wide spectrum of clinical severity, ranging from benign to life threatening. This wide range of clinical presentation, the relationship with different pathologies and comorbidities, the varied evolution and the scant knowledge of aetiology, often lead to difficulty in recognizing PI and frequently it is an occasional finding. In this paper the Authors present three cases with very different age and clinical onset, all related to chronic or sub-acute bronchopneumopathy causing dyspnoea and persistent cough in the previous months. The PI, associated with these pathologies, can be caused by a mechanical pressure gradient, but also by a currently unknown inflammatory mechanism.

Keywords: Asthma; Autoimmune Disease; Bronchopneumopathy; COPD; Intestinal pneumatosis

Introduction

Intestinal Pneumatosis is a rare disease with an etiology and pathogenesis which is often considered idiopathic [7]. The clinical presentation is variable. In this paper the Authors, other than a review of the recent literature, present three well documented and homogeneous cases with different age and clinical onset: a case of occasional asymptomatic finding, a case with chronic symptoms and a third case presenting with a picture of acute peritonitis. All of our patients have been documented as having chronic or sub-acute bronchopneumopathy causing dyspnoea and persistent cough in the previous months.

Materials and Methods

A comprehensive literature search was performed in order to find studies regarding PI and bronchopneumopathy. We used the uppercase boolean operators AND to combine and search terms related only to “Pneumatosis AND Bronchopneumopathy/COPD” and “Pneumatosis AND Asthma” in PubMed. Special emphasis was placed on papers related to the diagnosis and management of human patients in the last 5 years.

The cases we are presenting in this paper were collected by the internal database of our Surgery Department, and we selected patients with a radiological diagnosis (CT-scan) of PI.

Case 1

A 51-year-old male patient, in good habitual health, came to our attention with abdominal pain. For several hours before his

arrival to hospital, he reported pain in the right side of the abdomen (VAS 9/10), migrating to the suprapubic region without nausea, vomiting or diarrhoea. The patient, who denied recent colonoscopy or other abdominal diagnostic tests, was hemodynamically stable, afebrile and, on breath sounds were present throughout without added sounds. The abdomen was distended and painful in the lower right abdomen and suprapubic region, without signs of peritonism and with a clearly reduced peristalsis.

The patient reported right knee surgery and chronic cough. A Chest-CT carried out 6 months previously to determine the cause of a chronic cough showed a small nodular benign opacity of approximately 4.5 mm in the anterior segment of the right upper lobe.

A recent abdomen-CT showed 1 mm kidney stone at the right papilla, with minimal distension of the renal pelvis, the presence of parietal air in ascending-colon and at the splenic flexure (Figure 1). Blood tests showed elevated white cell level count (Leucocytes 17.9 $x10^9$/L with Neutrophils 15.2 $x10^9$/L) and microhematuria with rare bacteria in the urine.

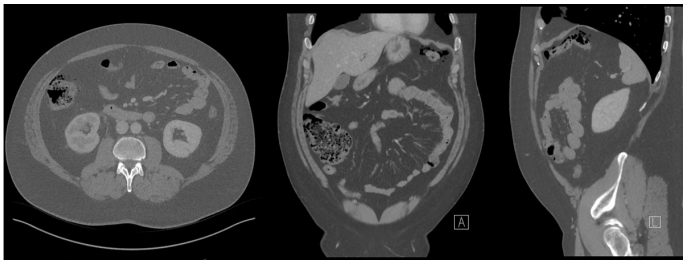

Figure 1: Case 1: F.R., 51-year-old male. 2° degree hydronephrosis of the right kidney from micro calculus at the urethral meatus level. Parietal air in the ascending colon and in the splenic flexure. Absence of free liquid.

The clinical presentation was interpreted as right ureterolithiasis. The patient was hospitalized for 2 days and discharged in good general condition with complete regression of the symptoms.

Studying the medical notes, carrying out a targeted respiratory history with the patient and with his Family Physician, we determined the chronic cough had been present for months and the most frequent causes of the respiratory symptoms had been excluded. At the time of hospital admission for ureterolithiasis the diagnostic process for chronic cough was complete and no obvious cause had been discovered. After 3 months of follow-up, all symptoms were completely resolved.

Case 2

A 67-years-old female patient came to our attention with widespread abdominal pain present for several weeks. She took Methotrexate and Prednisone for rheumatoid arthritis, and Fluticasone for asthma. She denied constipation or diarrhea. She reported abdominal pain bilaterally at the flankes and posteriorly at the left lung base. At past medical history there were asthma with poly-allergies, rheumatoid arthritis (anti-CCP positive), Sjögren syndrome with sicca syndrome, fibromyalgia and hypercholesterolemia.

Blood and urine tests were normal, without an inflammatory syndrome.

Complete abdomen-CT (Figure 2) documented PI (already present at a previous abdomen-CT carried out 16 months before) and a reduction of the colonic diameter associated with a stenotic effect.

The patient underwent laparoscopy (Figure 3), which was converted to a laparotomy for left hemicolectomy (resection of the transverse and descending-colon). Surveillance was performed in the intensive care. The progressive reintroduction of food was well tolerated, with a good recovery of intestinal function. The clinical evolution was good and the patient was discharged after 11 days in good health.

The histological examination of the surgical resection documented a Pneumatosis Intestinalis of the left colon wall, extending to the mesenteric adipose tissue (Figure 4). At one-month and 3-month follow-up, the patient showed good healing of the surgical wound. Intestinal function was normal without any further symptoms.

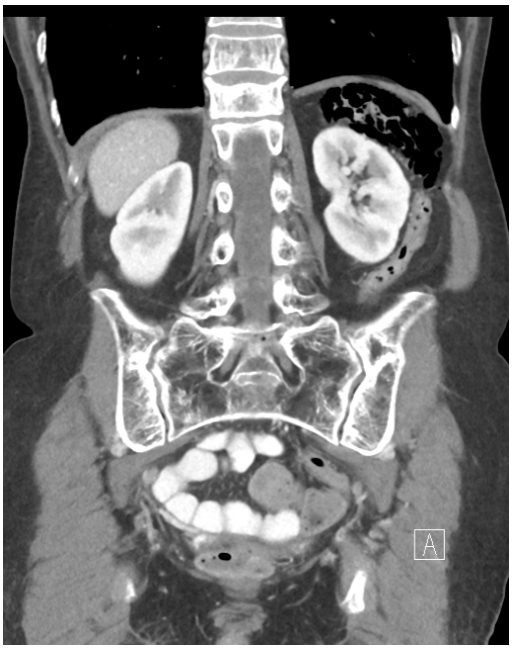

Figure 2: Case 2: 67-year-old female. Pneumatosis Intestinalis of the perivisceral fat of the left colic flexure with stenotic effect. No free air in the other abdominal quadrants.

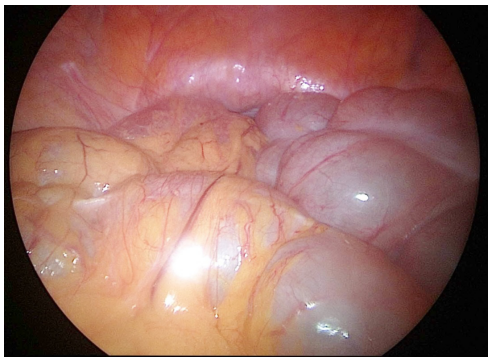

Figure 3: Case 2: 67-year-old female. Intraoperative view of laparoscopy: Pneumatosis Intestinalis of the transverse colon.

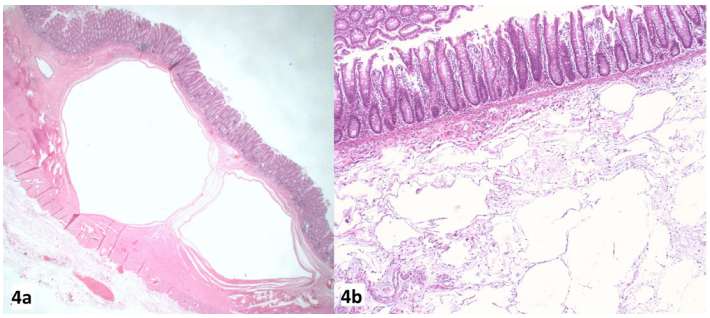

Figure 4: Case 2: 67-year-old female. 10x magnification (4a) and 50x magnification (4b) of Anatomical surgical resection. Pneumatosis Intestinalis of the left colic wall, extended to the mesenteric adipose tissue.

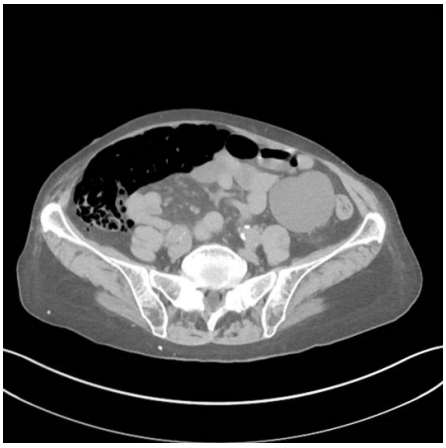

Figure 5: Case 3: 77-year-old female. Intestinal perforation at the level of the cecum/ascending colon with Pneumatosis Intestinalis.

Case 3

A 77-year-old female patient came to our attention with abdominal pain and fever at 40 ˚C. For two days she complained of abdominal pain localized in hypo-mesogastrium, without nausea, vomiting or diarrhoea. The family physician prescribed Ciprofloxacin 250 mg 2/day and Metamizole. Despite the therapy, the patient suffered an episode of double incontinence with loose stools.

Past medical history included a previous oesophageal carcinoma (T2 N0 M0) and a previous invasive ductal carcinoma of the left breast. The patient was anticoagulated with Acenocoumarol for atrial fibrillation, Methotrexate and Prednisone for systemic scleroderma and with Budesonide for asthma.

On clinical examination peristalsis was normal and present throughout with distension of the right abdominal quadrants and pain on deep palpation mainly in the hypogastrium. No signs of peritonism were present. Examination of the chest was normal, with reduced breath sounds at the lung bases.

Blood and urine tests showed an inflammatory syndrome (PCR 204 mg/L; Leucocytes 10.7 x10^9/L). On suspicion of an acute abdomen, an erect chest X-ray and an abdomen-CT were performed (Figure 5) which showed a right lung base infiltrate and an intestinal perforation at the level of the cecum/ascending colon.

The patient underwent a right hemicolectomy and was discharged in good general condition after 17 days of hospitalization. The histological examination of the surgical resection (Figure 6) confirmed a Pneumatosis Intestinalis of the colon. At one-month and 3-month follow-up, the patient showed healing of the surgical wound. Intestinal function was normal.

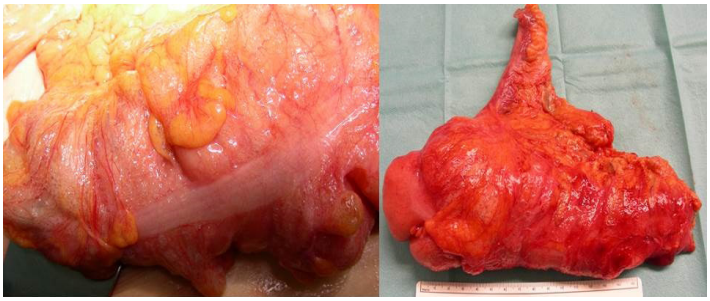

Figure 6: Case 3: 77-year-old female. Anatomical surgical resection with Pneumatosis Intestinalis of the colon.

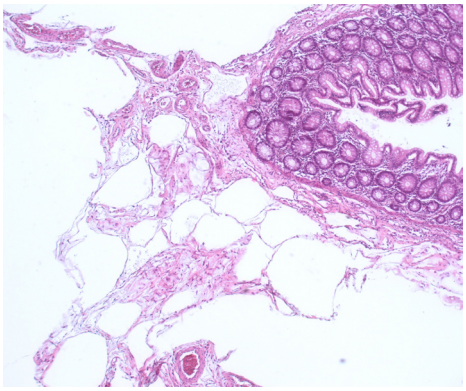

Figure 7: Case 3: 77-year-old female. 50x magnification of Anatomical surgical resection. Pneumatosis intestinalis of the ascending colon**.**

Conclusion

The literature review showed few articles focused on COPD and PI [7-10], but only one study stressed the possible relationship between these two pathologies [10] suggesting, as a possible aetiopathogenic mechanism of PI, the alveolar air leakage secondary to high airway pressure due to chronic obstructive pulmonary disease. PI has also been described in patients with connective tissue disorders such as Sjögren Syndrome and systemic sclerosis [6], or with several states of immunodeficiency [2-5].

The cases described in our series (Table 1), although very heterogeneous, have in common chronic respiratory diseases or prolonged cough and, in two of them, an autoimmune disease was also present with a concomitant steroid therapy.-

	SEX	AGE	COMORBIDITIES	RELEVANT LABORATORY RESULTS	LENGTH OF DISEASE DEVELOPMENT	ONSET
Case 1 (F.R.)	M	53	- Chronic Cough [9] - Hepatic Steatosis	Leucocytes 17.9 x 10^9/L (Neutrophils 15.2 x10^9/L)	Undetermined due to the occasional finding and asymptomatic nature	Occasional asymptomatic finding
Case 2(R.M.)	F	67	- Asthma - Sjögren syndrome - Rheumatoid Arthritis anti-CCP positive	Not acute Inflammatory Syndrome	3 Months	Chronic and Diffuse Abdominal Pain
Case 3 (V.C.A.)	F	77	- Asthma - Systemic Scleroderma - Bacteraemia - Squamous-cell Carcinoma (“en-block” Gastrectomy, Chemotherapy, Radiotherapy) - Breast Invasive Ductal Carcinoma - Osteoporosis	Inflammatory Syndrome (PCR 204 mg/L; Leucocytes 10.7 x 10^9/L) Potassium 2.4 mmol/L Lactate 2.5 mmol/L	2 Days	Acute Abdominal Pain

Table 1: Case series summary table.

Respiratory disorders can cause increased pressure at the level of the respiratory tract and more easily induce air passage through the mediastinum or diaphragm causing PI. On the other hand, the simultaneous autoimmune pathology and the steroid treatment may have further subverted the consistency of the connective tissue and contributed to the diffusion of the air.

Disclosure

We confirm that there are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome.

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